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INTRODUCTION

The term scleroderma literally means hard skin. But scleroderma (also known as systemic sclerosis, or SSc) is much more than a skin disorder.

While nearly all persons with systemic sclerosis have issues with their skin, particularly excessive scarring, swollen, tight, or hard skin on their fingers, they also have organ system involvement. Many have tight, swollen, or hard skin in other body areas as well, particularly the face and the arms. Additionally, several other types of organ system involvement are also characteristic of systemic sclerosis, primarily the vascular and the immune systems. For example, blood vessels are frequently affected (vasculopathy), leading to spasmodic color changes (red, white, or blue) brought on by cold exposure. This is commonly referred to as Raynaud phenomenon and it occurs in the fingers of about 90 percent of persons with systemic sclerosis.

One of the most common manifestations of immune involvement is the presence of abnormal levels of autoimmune antibodies to the nucleus of one’s own cells (anti-nuclear antibodies or ANA) that are seen in nearly everyone with systemic sclerosis. These three clinical features—excessive fibrosis (scarring), vasculopathy, and autoimmunity—appear to underlie the processes that result in the different manifestations that characterize systemic sclerosis.

TYPES OF SYSTEMIC SCLEROSIS

For reasons that are not clear, the normal protein collagen is deposited in the skin of patients with systemic sclerosis in higher amounts than in the skin of people who are disease-free. This process makes the skin thick and tough. In some persons, the excess collagen deposits involve only the fingers and possibly the face or hands. In others, the deposits can be found in areas all over the body. Furthermore, systemic sclerosis patients are placed into two additional subgroups based on the extent of skin involvement. These subgroups are diffuse cutaneous scleroderma and limited cutaneous scleroderma.
Diffuse cutaneous scleroderma

This subgroup is characterized by thick or tight skin on the arms, above and below the elbows, and frequently on the legs, above and below the knees, with or without involvement of the face. The skin on the torso (chest and abdomen) is frequently tight, thick, or hard. The thickening of skin often progresses rapidly and is very bothersome, leading to thickening of skin in many areas all over the body in a short period of time (weeks to months). It is not uncommon for the thickening process in the skin in diffuse cutaneous scleroderma to continue for one to three years before the thickening process slows down and levels off. After one to two years of stability in the thickness, the thickening usually begins to recede and the skin begins to thin or soften.

Limited cutaneous scleroderma

People with this form of the disease have thick, tight, or hard skin on areas below, but not above, the elbows and knees, with or without involvement of the face. Thickening of skin frequently develops gradually and is relatively unobtrusive. When measured repeatedly over time, the skin score in patients with limited cutaneous scleroderma is usually small and changes very little, even over many years.

What does this division into limited and diffuse cutaneous scleroderma tell us?

Both subgroups are part of a more general disorder called systemic sclerosis and, as such, both subgroups...
and limited cutaneous scleroderma. Sclerosis patients are placed into two additional areas all over the body. Furthermore, systemic collagen deposits involve only the fingers and possibly the face or hands. In others, the deposits can be found in areas all over the body. For reasons that are not clear, the normal protein collagen is deposited in the skin of patients with collagen deposits. This process makes skin score in patients with limited cutaneous scleroderma tell us? What does this division into limited and diffuse cutaneous scleroderma tell us? Both subgroups are part of a more general disorder called systemic sclerosis and, as such, both subgroups share some features in common, including:

- Raynaud Phenomenon. This occurs in about 90 percent of patients with systemic sclerosis.
- Heartburn and other esophagus problems (particularly trouble swallowing foods).
- Skin sores are common, primarily on the fingers. Some patients develop sores on the skin of the wrists, elbows, or ankles.
- Abdominal grumblings that can include feeling “full” after eating only a small of amount of food, “bloating” of the belly after eating, swelling of the abdomen, particularly after eating, constipation, or diarrhea.
- About 10–15 percent of patients may develop severe lung fibrosis, leading to shortness of breath.
- About 10–20 percent of patients may develop pulmonary hypertension, which is high blood pressure in the arteries that supply the lungs.

While the above-listed characteristics are those that each subgroup has in common, there are features that are seen more commonly in one of the two subgroups.

### Features seen more commonly in diffuse cutaneous systemic sclerosis:

- Kidney failure in about 15–20 percent of patients, fortunately for which there are treatments that can preserve kidney function and prolong life for many, if treated early.
- Approximately 10 percent of patients will experience some level of heart involvement, including fluid around the heart, heart rhythm disturbances symptomatic enough to require treatment, and possibly heart failure.
- Musculoskeletal aches and pains, decreased motion of some joints (fingers, wrists, elbows, shoulders, and occasionally knees) and declines in hand function that can result in disability are fairly frequent.

## TYPICAL ORGAN INVOLVEMENT

### Skin

The skin is the largest organ in the body. Its disadvantage is that it is spread over the surface of skin.
the body and not located in one area, like the heart or the kidneys where it can be easily examined and measured. Because it is spread all over, new methods for measuring the degree of thickness over the body have had to be developed. The most frequently used assessment is a technique called skin scoring. Scoring is done by a medical professional who feels and pinches the skin in 17 body areas and assigns a number to the thickness of skin in each of the areas on a 0–3 scale (zero being normal and 3 being very thickened). The scores in all 17 areas are added together to give the skin score (the range of scores is 0–51). Using this technique, skin scores of patients with limited and diffuse cutaneous scleroderma have been documented to change over time in fairly characteristic ways.

**Lungs**

The vast majority of persons with systemic sclerosis will develop some scarring in the lungs, usually seen on a high-resolution chest computed tomography, or CATH scan. The majority of these persons will never develop serious lung involvement. However, in about 40 percent of patients, there will be some measurable decline in lung function. The vital capacity (a measure of how much air your lungs can move in and out in one deep breath) may get smaller because scars in the lung tissue make the lung stiffer. The good news is that there have been encouraging treatment studies recently that suggest some immunosuppressive treatments (i.e. cyclophosphamide) may slow damage to the lungs. More research is ongoing that is looking into this and other treatment strategies to slow lung damage.

**Kidneys**

Kidney failure is a severe complication that occurs mainly in persons with diffuse cutaneous scleroderma who have had systemic sclerosis for less than five years. Problems begin with a decline in blood flow to the kidneys for reasons that doctors do not understand. This then triggers the release of hormones, which, unfortunately, results in elevations in blood pressure and further reductions in blood flow through the kidneys. This is commonly called
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**TYPICAL ORGAN INVOLVEMENT**

- About 10–20 percent of patients may develop abdominal grumblings that can include feeling “bloating” of the belly after eating, swelling of the “full” after eating only a small of amount of food, “gassy” or “wind” stomach, or “gas” in the lower abdomen.
- Heartburn and other esophagus problems
- Skin sores are common, primarily on the fingers.
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**As a member of the Scleroderma Foundation, you will receive:**

- Our quarterly magazine, the “Scleroderma VOICE.” The magazine includes updates on the latest scleroderma research and treatments, positive and uplifting stories from patients living with the disease; and tips about how to manage living with scleroderma.
- Information and educational offerings from your local chapter.
- Discounted registration fees to the annual National Patient Education Conference.

Please consider joining the Scleroderma Foundation today. A membership form is attached on the reverse side of this panel.
Our Three-Fold Mission Is Support, Education and Research

SYSTEMIC SCLEROSIS: Diffuse and Limited
May 2015

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About 80–90 percent of persons with systemic sclerosis can be shown to have a “lazy” muscle in their esophagus. This can lead to heartburn and the sensation that food “sticks” in the chest part way down. Unfortunately, the muscle in the stomach and in the rest of the intestine can also get lazy. This can lead to a multitude of symptoms including filling up too fast after eating small amounts of food, a sensation of “bloating” or swelling in the belly after eating, chronic diarrhea or chronic constipation. Unfortunately many of these symptoms are not specific to any particular area of the gut and it may take a doctor and patient considerable time to try to sort out which area of the gut is causing the problem and which area needs treatment.

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**Support:** To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.

**Education:** To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.

**Research:** To stimulate and support research to improve treatment and ultimately find the cause of and cure for scleroderma and related diseases.

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